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ABSTRACT

Presented is an overview of hearing, speech and language research being sponsored by the National Institute of Neurological Diseases and Stroke (NINDS). Described is research in such areas as infant audiological screening, auditory prostheses, larynx surgery, and developmental dysphasia. (LS)

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The NINDS Hearing, Speech, and Language Research Program

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SPECIAL REPORT: HEARING, SPEECH, AND LANGUAGE

More than 20 million Americans have a speech, hearing, or language problem during some period of their life. The National Institutes of Health conducts and supports research to increase understanding of human communication and its disorders and to prevent these problems. Sharing this effort are the National Institute of Neurological Diseases and Stroke (NINDS), the National Institute of Child Health and Human Development (NICHD), the National Institute of Dental Research (NIDR), and the National Institute of Environmental Health Sciences (NIEHS).

The NINDS program in communicative disorders has 132 research projects, including five out-patient clinical research centers, seven contracts, 55 training grants to universities and medical centers, and five multidisciplinary centers. In addition, the Institute this year created a new Laboratory of Neuro-otolaryngology to study the biochemical, anatomic, and physiologic processes of the auditory system, basic to developing better methods of preventing and treating auditory disorders.

NINDS also has a Communicative Disorders Section in the Collaborative and Field Research Program which is expanding its directed research to target on new problem areas, particularly those which do not receive funding from other Federal agencies. These areas include improved objective identification of young hearing-impaired children; improved measurement, treatment and prevention of speech and language disorders among adults who have experienced stroke or head trauma; effects of noise on children; noise and speech communication; and improved ways to assess and treat language communicative disorders which are not apparently attributable to sensory impairments. The Section is currently supporting research on utilizing wearable master hearing aids and on a study of the extent to which deaf persons are able to integrate information obtained by lip reading with additional information derived from an electronic analysis of the acoustic speech wave form.

To make research information more readily available to scientists and the public, the Institute has supported an Information Center for Hearing, Speech, and Disorders of Human Communication, located at The Johns Hopkins University Hospital in Baltimore. The Center has sponsored several workshops on communicative processes and problems, has published biblioprofiles and state-of-the-art reports on communicative disorders, and has published a directory of organizations and publications in the hearing and speech field.

HEARING

Hearing Disorders

The ultimate goal of hearing research is prevention, but because the nature of the hearing mechanism is so complex, many auditory problems have eluded amelioration or cure. The answers should come, however, with an increasingly precise understanding of anatomic, physiologic and biochemical processes. Much of the NINDS emphasis focuses on these basic functions. Concurrently, support is given clinical studies on improved treatment for persons afflicted with hearing problems.

One of the targets for prevention is hereditary deafness. Recent studies indicate that approximately 40 percent of deafness present at birth is hereditary. More than 40 types have been identified, most of them resulting from faults in five or six genes. Two types: (1) recessive microtia (abnormally small ears) and hearing loss; and (2) recessive achalasia (tense gastrointestinal muscles), piebaldism (absence of pigment in skin and hair) and hearing loss were described within the past year by NINDS grantees at The Johns Hopkins University. Better understanding of hereditary deafness can aid counseling of prospective parents.

Another 10 percent of congenital deafness has resulted from prenatal rubella infection. The unique data collected from the NINDS collaborative perinatal study of more than 50,000 women and their offspring which was in progress during the rubella pandemic of 1964 have provided unique information on the correlation of rubella infection and deafness. Studies indicate that about half of the infants whose mothers had rubella during the first three months of pregnancy have deafness. Of these, approximately half also have impaired function of vestibular structures (part of the inner ear concerned with equilibrium which connects to the cochlea, the organ of hearing). Temporal bone studies by NINDS grantees at The Johns Hopkins University have shown varying degrees of vestibular problems, including defects of the cochlear duct and saccule which is sensitive to the pull of gravity. Fortunately, the recently developed rubella vaccine has sharply reduced the incidence of rubella-caused defects in babies.

These data, plus recent advances in virology techniques, can now bolster understanding of the effects of a maternal virus infection on the fetus. Learning how other viruses are implicated in congenital deafness should be of help in improving methods of prevention.

Although increased understanding is leading to better methods of prevention, there is still a great lack of knowledge in this area. Therefore, early identification of infants born deaf or with a hearing loss is vital since hearing problems can best be managed when they are discovered early. Physicians have relied on using a high risk register to identify infants with a high probability of deafness based on family history.

Screening procedures for all newborns have not proved successful because there has been no reliable test. This year, however, an exciting new apparatus for detecting a continuous lack of response to auditory stimulus has been introduced by an NINDS grantee at Stanford University. The new apparatus, called a "crib-o-gram," consists of a transducer mounted on a crib which can detect infant movement in the crib and record any changes when a test sound is turned on. Criteria for "passing" and "failing" the test have been developed through a control study of 5,000 babies. Infants are considered to hear if their response rates are better than 20 percent, or if there are two distinct arousal responses within two seconds after the test sound. The equipment is capable of testing 6,000 babies throughout the day (although the actual number would be dependent on the number of cribs available). Results are tabulated by computer.

In another contract-supported study, simple test devices suitable for home use by parents are being evaluated as a means of early detection of deafness in very young infants—an approach that would permit large-scale screening.

NINDS-supported researchers at the University of Chicago have developed a research technique for obtaining an objective test of hearing function. It may become useful in testing small children and hearing-inpaired adults. Called electrocochleography, it consists of measuring electrical impulses produced in the cochlea (the organ of hearing located in the inner ear) as they move along various nerve pathways from the cochlea to the brain's cortex. The technique involves passing a fine wire electrode through the eardrum to the promontory of the middle ear, and deriving information on the frequencies from different regions of the cochlea. Results are computerized and emerge as patterns. Conventional audiometry (quantitative and qualitative evaluation of hearing using an instrument known as an audiometer) measures excitation of the entire cochlea, but does not provide information on such specific frequency regions.

These tests, plus knowledge of a family history of hereditary deafness, are helping to identify early those infants with a hearing problem. But, hereditary deafness is not confined to infants. In fact, there are 30 types of hereditary deafness for which onset is delayed. The most common of these is otosclerosis, caused by a dominant gene with variable penetrance. Surgery for otosclerosis, one of the most important advances in the field, usually can restore hearing in most patients. In otosclerosis, growth of spongy bone in the middle ear immobilizes a small middle ear bone, the stapes, that normally vibrates and conducts sound to the inner ear. Surgically removing and replacing the stapes is reported to be effective in more than 90 percent of the patients, and the procedure has become increasingly safe with development of refined microsurgical techniques.

Research on preventing otosclerosis includes studying the initial defects. Electron microscopy and tissue studies show that changes in otosclerotic areas

may be due to abnormalities in bone-producing cells (osteocytes) or in certain white blood cells which destroy bone (macrophages). Future biochemical studies may lead to methods of preventing these changes. In some studies, sodium fluoride has been demonstrated to arrest the otosclerotic process and may offer some hope for a successful drug treatment.

Another condition in which surgery can dramatically prevent not only hearing loss but even death is acoustic neuroma. Safe removal of these tumors growing on the sheath covering the acoustic nerve is possible because of striking advances in methods of early diagnosis (sophisticated audiological tests and cerebellopontine angle myelography) and excellent microsurgical techniques. These techniques, coupled with prompt management of postoperative complications, have ushered treatment into a new era, far removed from the days in which high operative mortality rates militated against surgical removal of such tumors.

Surgery is also being used, though sparingly, for some persons with Meniere's disease. This disorder, characterized by hearing loss, tinnitus (ringing in the ears) and vertigo, often will respond to medical measures including diet regulation, control of allergies, and drugs for treating the symptoms. In some persons for whom these measures are not adequate, cryosurgery and ultrasound are occasionally used in the inner ear, destroying the end organ of balance, the vestibule, but preserving cochlear (hearing) function. However, even when surgery is performed, symptoms often recur with a slow progression of hearing loss. The final answer to Meniere's disease is expected to come with increased understanding of inner ear (vestibular) physiology.

More research is also needed on finding beneficial drugs for persons with Meniere's disease. A recent study of the effects of streptomycin on the sensory cells of the vestibular system in animals indicated that the drug could selectively destroy the vestibular apparatus with no damage to the cochlea. This would leave hearing intact. More research is needed, however, to determine the possibility of using this therapy on humans.

Streptomycin can be toxic to persons with normal hearing. It is, in fact, only one of several drugs found to affect hearing. During the past several years, the toxic doses and site of injury were defined, some by NINDS grantees, for dihydrostreptomycin, neomycin, kanamycin and ethacrynic acid. To alert practicing physicians, and acquaint researchers with current knowledge on these and other drugs which may be toxic to hearing, the NINDS-supported Information Center for Hearing, Speech, and Disorders of Human Communication this year published a book entitled "Index-Handbook of Ototoxic Agents, 1966-1971."

Still another contributing factor to hearing loss can be otitis media, an inflammation of the middle ear space. Infectious otitis media commonly develops from infection in the nose or nasopharynx. It can cause up to a 35-40

decibel hearing loss if left untreated, but is easily treated with antimicrobial agents and oral or topical decongestants. Serous otitis media results from fluid collecting in the middle ear, often after prolonged blockage of the eustachian tube. Serous otitis media, usually unaccompanied by pain, can also cause hearing losses which often go undetected for considerable periods of time. Although treatment with oral and topical decongestants, enzymes, and occasionally antibiotics is often effective, loss may reoccur with eustachian tube malfunction. Current research, therefore, is centered on eustachian tube function and management of tubal disease.

Even when no outside direct causes of hearing loss are evident, with increasing age there is inevitably a variable sensori-neural and discrimination hearing loss. Hearing loss accompanying the aging process remains one of the most prevalent and as yet, incurable hearing problem. Scientists have identified pathological changes in the temporal bone, including degeneration of certain inner ear structures. Other studies have indicated that diseases of the blood vessels may be responsible for degeneration seen in presbycusis (loss of hearing of high pitched tones). Studies by NINDS-supported researchers at The Johns Hopkins University, however, indicate that other factors may be involved. They demonstrated a loss in volume but not in number of auditory neurons. A more thorough understanding of auditory anatomy and physiology may provide clues to understanding, and then staving off the occurrence of these unknown variables.

Recent work in auditory physiology is helping to unravel the complex interconnected systems involved in hearing. Tremendous advances in the study of the mechanics of the auditory system have been made with the use of new ultrasensitive techniques. The way information is coded in many nerve fibers is now relatively well understood. Studies indicate that most of these neurons have the same properties so they can be measured in the same way. In the inner ear, however, there are diverse coding properties. Researchers feel that as techniques for electrophysiological recording from animals are employed and improved, a more exact understanding of the central auditory pathways can be gained.

Limited understanding of the coding process in the inner ear and technical difficulties present formidable obstacles to artificially generating the neural information which normally enters the brain via the auditory nerve. Many scientists believe the key to a successful implanted hearing device will be discovering how the normal ear encodes auditory information and how it is utilized by the central nervous system.

Researchers in several universities, medical centers, and private industry are working on prosthetic devices for the deaf. This may be one of the most exciting areas of auditory research, but it is also one of the most frustrating. Since development and implantation of prosthetic devices involve working

with the brain and nervous system at their most complex level, it is one of the most difficult areas in auditory research.

One type of device commonly referred to as an "implantable hearing aid" is a very small electromechanical transducer which causes a mechanical vibration in the inner ear. This device would be most helpful for persons with conductive hearing loss (occurring when sound waves are not transmitted adequately to the inner ear). It might also be able to reduce distortion for some persons with sensori-neural loss. Initial mechanical design of such an aid has been completed. However, other critical problems, including that of foreign-body reaction (scarring), remain to be solved.

Another type of sensory prosthesis under study by NINDS researchers is one which would directly stimulate the auditory nerve, the main nerve of hearing. Patients participating in early prosthesis trials were able to discriminate pitch and amplitude, but were not able to perceive complex tones and speech. Understanding of the coding process would greatly enhance chances for success with this type of prosthesis.

Other researchers are considering direct stimulation of the auditory portions of the brain. Three years experience in the Institute's collaborative program to determine the feasibility of several neural prostheses has provided scientists with valuable information that may have application for development of an auditory prosthesis. For instance scientists have found that different parts of the brain respond to different tones. It may eventually be possible by stimulating appropriately placed electrode arrays on brain tissue to enable deaf persons to discriminate speech.

While auditory prosthesis research still faces difficult obstacles, research on improving existing aids for the hard of hearing is helping increase the quality and the wearer's efficiency in using residual hearing. Presently, sensory devices are of two kinds: acoustic hearing aids which are wearable sound amplifiers; and sensory coding aids, which convert sound so that it may be sensed through vision or touch. Research on acoustic hearing aids is designed to maximize their usefulness and to gain knowledge of how best to fit and adjust the aids to the wearer's individual needs. The Institute supports work on this at Northwestern University and the Central Institute for the Deaf.

This year the Institute has had developed, under contract, a master hearing aid which will enable scientists to determine methods for tailoring hearing aids to be of greatest aid to the individual wearers. Research on improving fidelity, increasing ruggedness, and reducing distortion would increase aid efficiency.

Sensory coding aids are in use in prototype forms such as visual speech indicators for speech training. One visual aid to speech reception is mounted on eyeglasses. An NINDS contract with Gallaudet College involves prelim-

inary evaluation of an automatic system for separating out speech sounds and placing them in categories in which the deaf person can tell the sounds apart through lipreading. The system will present its output to the user in either a visual or tactile code.

Another contributing factor to hearing loss is noise. At present the only method of dealing with hearing loss resulting from excessive noise is prevention. Research supported by NINDS at a number of universities and research centers is providing an understanding of the basic physiological structures and functions that are adversely affected by exposure t noise. Some of these studies are providing a quantitative correlation between noise dosage accumulated over a person's lifetime and impaired hearing.

A recent laboratory study indicates that it is to be expected that even after repeated exposures, approximately 25 percent of people living in neighborhoods near airports and highways will be awakened by each intense airplane or truck noise. It has been established that permanent hearing loss can be produced by loud noise, particularly industrial, or rock-and-roll music. One researcher found that 10 percent of persons listening for two hours to rock-and-roll music at 110 decibels (a unit of sound) would have permanent hearing loss. Damage was found in the organs of Corti of guinea pigs exposed to noise from one minute to 24 hours at 100 to 138 decibels by researchers in Ohio. In addition, there was disintegration and loss of spiral ganglion cells which many scientists feel may be responsible for the permanent hearing loss.

SPEECH

About 10 million people in this country have a speech disorder which significantly interferes with communication; half of these are children, and another sizeable proportion appears in the older age groups.

Speech Disorders

Normally, speech activity originates and is organized in the brain's cerebral cortex. Information flows out through particular nerve tracts to the muscles involved in producing speech. Then there is a sensory flow back to the central nervous system, probably indicating the position of all the structures related to speech, allowing other brain centers to project the necessary nerve impulses for a smooth flow of motor activity. An interruption in this process often accompanies neurological disorders such as cerebral palsy, stroke, multiple sclerosis, Parkinson's disease, dystonia and many others.

Speech disorders resulting from disturbances in muscular control of the speech mechanism caused by damage to the central and peripheral nervous system are known as dysarthrias. They are believed to be the result of disruption at the lowest level of output. This implies that disturbances within

the nervous system can affect any or all aspects of speech production—respiration, phonation, articulation, and resonation. During the past few years, several studies have been made on the perceptual, physiologic, acoustic and rehabilitative aspects of dysarthria.

Recent studies of patients with multiple sclerosis indicated that less than half had significant speech deviation. But a variety of deviant speech dimensions was noted in those who were dysarthric. The severity was related to the extent and severity of motor-nerve involvement. Although studies such as these suggest that dysarthria is an impairment of motor control, other research indicates a high probability of sensory deficit as well, since sensory and motor pathways are in close proximity. For instance, results of a recent study showed significant sensory and perceptual deficits on tests of oral form identification and two-point discrimination of the tongue. Using lateral cineradiography (X-ray motion pictures) researchers were able to describe the nature of the disturbed tongue movements during swallowing and speech in patients with Parkinson's disease. Their results are defining the pattern and sequence of deterioration in vocal tract control in Parkinson's disease and can be applied to other progressive neurologic diseases. Characteristically, patients with Parkinson's disease have tension and rigidity of the muscles directly affecting speech. NINDS-supported scientists utilized bioelectric feedback to reduce lip tension in a patient with Parkinson's disease, removing undesirable lip retraction.

Speech disorders also arise, of course, as a consequence of laryngectomy operations for cancer of the larynx which strikes approximately 6,000 persons each year. Dramatic advances have been made in maintaining or restoring speech function following surgery for removal of the larynx which contains the vocal cords. Improved methods of detection are enabling doctors to make an early diagnosis, before the damage is too widespread. In these cases, conservation surgery is being used—that is, removing only the cancerous parts of the larynx, which provides the best prognosis for a functioning larynx. Following recent approval by the Food and Drug Administration of using teflon (suspended in glycerin) for injections to strengthen remaining tissue, restoration of normal speech and the ability to swallow have been made possible in a large percentage of patients.

An exciting advance in larynx surgery this year was made by scientists in Massachusetts. They used a carbon dioxide laser integrated with an operating microscope to selectively evaporate predetermined amounts of abnormal vocal cord tissue through a laryngoscope. Removal was reported to be precise and rapid. Follow-up examinations indicate normal healing similar to that in clean surgical wounds.

A head and neck registry is being kept at Washington University in St. Louis to maintain statistics on the long-term survival rates and quality of

life of patients who have had the larynx removed. For those whose cancer is too widespread for conservation surgery, speech prostheses are being developed by several investigators, while others are studying in animals the possibility of reconstructing a normal larynx. Experimental prosthetic aids include a pneumatic device called the La Barge prosthesis developed by scientists in New York. It involves utilizing esophageal speech which is produced by swallowing air into the esophagus (food pipe) and expelling it with a burp to the mouth where the sound is modulated by the lips and tongue to produce sound. The La Barge prosthesis connects a fistula (hole) in the neck to the esophagus, so that the air used for breathing can also be used for esophageal speech by vibrating remaining tissue. This procedure, which has had limited use by human volunteers, is still in the investigative stage.

A second device, called the Tokoyo larynx, developed at Purdue University, helps to create sound by acting like a wind instrument, making a buzzing sound as air passes through it. The sound is carried through a tube to the mouth, where sounds are modified by the person's articulation.

A third device, called the Western Electric Hand Held Larynx, consists of a vibrator held next to the neck. It generates a pulse which is transmitted through neck tissue, and serves as a sound source. These devices are helping patients produce intelligible speech. However, it is hoped that research on the process of speech production, including frequency, duration, and intensity of speech sounds, will lead to better speech aids.

Computers are now being used by several NINDS investigators to provide analysis of the speech process. Other NINDS-supported researchers, at Washington University, St. Louis, and the University of Cincinnati, are working to reconstruct the larynx from natural tissue. Animal studies at Cincinnati, for instance, have shown that a functioning larynx in dogs can be made from their own throat tissues. Studies are now being made to see if this procedure can be successful in baboons, whose upright stance is similar to man's. Construction of the new larynx has been completed, and scientists are now attempting to open it and construct vocal cords.

LANGUAGE

Language, a formal system of signs and symbols commonly used for transmitting and understanding ideas, is usually accomplished through written or spoken words. Everyone must learn it or be at a disadvantage socially and educationally. Currently, it is estimated that more than three and one-half million children between the ages of four and seventeen have some degree of language disability.

Language Disorders

Disorders affecting language development can be present from birth, often the result of deafness, vocal paralysis, mental subnormality, early infantile autism and specific brain damage. Other children who do not suffer from any of these problems but still fail to develop normal language often have what is known as developmental dysphasia.

Language problems also involve children and adults as a result of brain trauma or stroke. Their condition is usually diagnosed as asphasia, the inability to either assign meaning to words, repeat them, or organize words into thoughts. Approaches to language rehabilitation generally follow one of four procedures. They may be based on: Language structure (building up to increased levels of structural complexity); language acquisition of the normal child; perception and cognition (subordinating concern for linguistic structures); and stimulus-response combinations.

Research on developmental dysphasia has centered on two approaches: search for evidence of early damage to the central nervous system; or for evidence of high-level auditory perceptive deficits. Recent evidence indicates that the latter is involved in dysphasia. NINDS-supported scientists at the Institute of Childhood Aphasia at Stanford University described auditory perceptual impairments, demonstrating that children with developmental dysphasia have difficulty sequencing sounds which they hear. Other research showed that persons with dysphasia have impaired ability to discriminate sounds which require rapid auditory analysis. The findings indicate that dysphasic children do not have the ability to perceive many important speech sounds and therefore fail to develop speech.

Continued research on speech sounds which are artificially slowed down by computer may yield important insight into the mechanisms involved, and may also be useful in therapy for children with dysphasia.

By contrast with the milder dysphasias, the term aphasia is usually reserved for a severe and devastating language disorder which produces anxiety, confusion and torment in its victims who once had normal speech and may have lost it as a result of stroke, brain tumor, or brain infections. Persons with aphasia usually have had a strong language base before onset of the disorder. They frequently have less severe language impairment, their problems are usually restricted to expressive abilities (rather than initial understanding), and the problem can usually be associated with specific brain damage. Aphasia can manifest itself in an individual's listening, speaking, reading, and writing abilities. NINDS-supported studies at the Boston Veteran's Hospital showed that not all aspects of the language processes are equally impaired, and persons with aphasia were able to perceive linguistic stress in the same way normal persons do. During the past few years, a test battery for determining the extent of aphasia and for predicting levels of rehabilitative success was

developed by NINDS-supported researchers at the University of Iowa. More research is needed to develop methods for testing and measuring language disorders, to determine the nature of the disturbances, and to devise more effective treatment and rehabilitation.

SHARING RESEARCH FINDINGS

The dissemination of research findings and their application are considered by NIH to be one of its major functions in promoting relevant and appropriate research.

The NINDS-supported Information Center has supported a number of workshops during the past few years. The proceedings of many of these have been published, including Vascular Disorders and Hearing Defects; Sensory Input in Hearing Impaired Children, and both Neuroanatomy and Physiology of the Auditory System. The Center has also had issued several publications including: Index-handbook of Ototoxic Agents, 1966-1971; Biblio-Profiles (including Homotransplantation, Auditory Physiology, Otitis Media, Surgical Treatment of Deafness, Viral Infection and Hearing, Neuroanatomy of Speech, and Rehabilitation of Language Disorders in Children); research bibliographies; and two directories, Information Sources in Hearing, Speech and Communication Disorders—Publications and Organizations.